# BEST AVAILABLE IMAGES

Defective images within this document are accurate representations of the original documents submitted by the applicant.

Defects in the images may include (but are not limited to):

- BLACK BORDERS
- TEXT CUT OFF AT TOP, BOTTOM OR SIDES
- FADED TEXT
- ILLEGIBLE TEXT
- SKEWED/SLANTED IMAGES
- COLORED PHOTOS
- BLACK OR VERY BLACK AND WHITE DARK PHOTOS
- GRAY SCALE DOCUMENTS

# IMAGES ARE BEST AVAILABLE COPY.

**As rescanning documents *will not* correct images, please do not report the images to the Image Problem Mailbox.**

uence Range: 1 to 6200

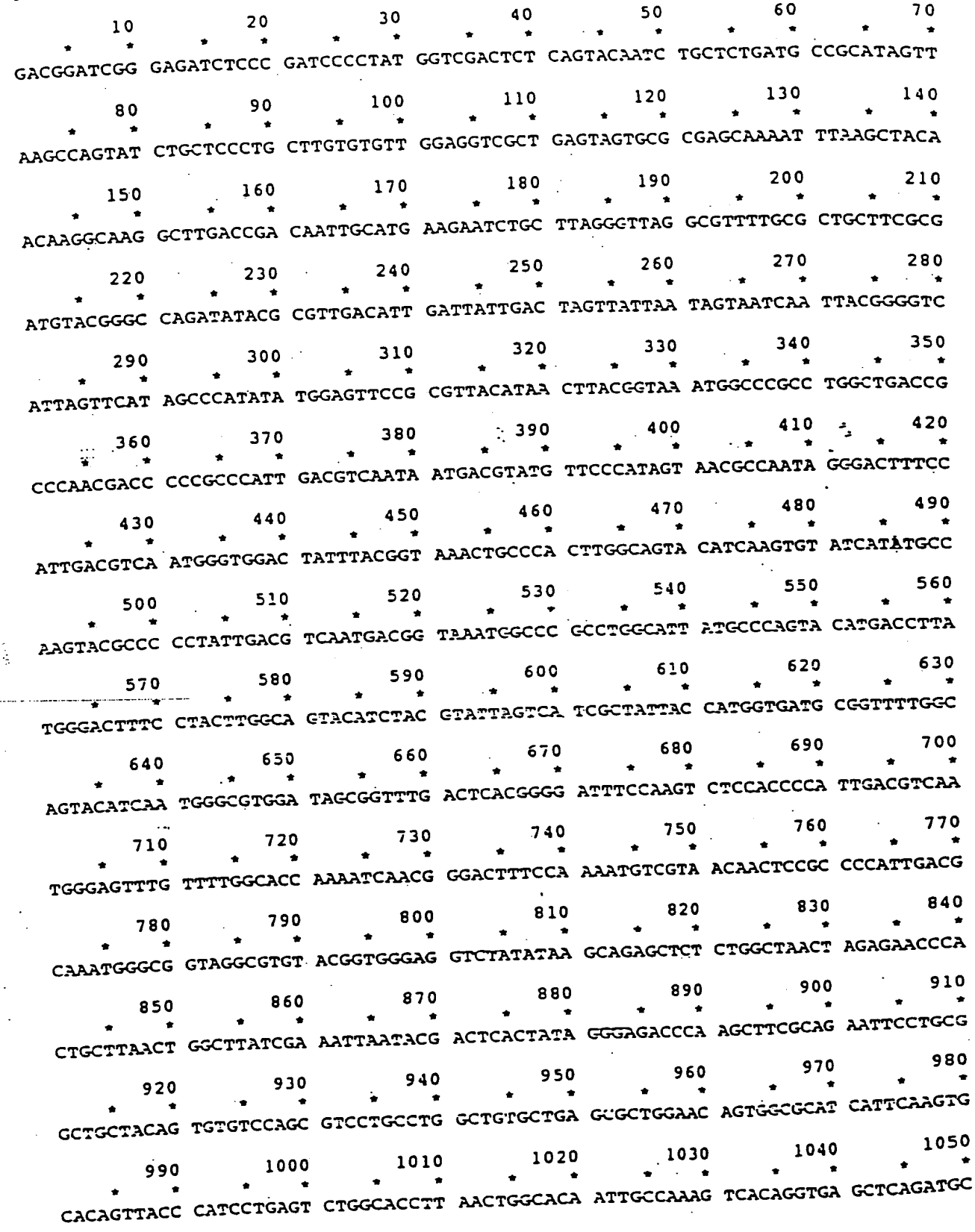



FIGURE 1

Homo sapiens

```
                                                                    1060        1070        1080        1090        1100        1110        1120
                                                                     *   *       *   *       *   *       *   *       *   *       *   *       *   *
ATACCAGGAC ATTGTATGAC GTTCCCTGCT CACATGCCTG CTTTCTTCCT ATAATACAGA TGCTCAACTA

      1130        1140        1150        1160        1170        1180        1190
       *   *       *   *       *   *       *   *       *   *       *   *       *   *
ACTGCTCATG TCCTTATATC ACAGAGGGAA ATTGGAGCTA TCTGAGGAAC TGCCCAGAAG GGAAGGGCAG

      1200        1210        1220        1230        1240        1250        1260
       *   *       *   *       *   *       *   *       *   *       *   *       *   *
AGGGGTCTTG CTCTCCTTGT CTGAGCCATA ACTCTTCTTT CTACCTTCCA GTGAACACCT TCCCACCCCA

      1270        1280        1290        1300        1310        1320        1330
       *   *       *   *       *   *       *   *       *   *       *   *       *   *
GGTCCACCTG CTACCGCCGC CGTCGGAGGA GCTGGCCCTG AATGAGCTCT TGTCCCTGAC ATGCCTGGTG

      1340        1350        1360        1370        1380        1390        1400
       *   *       *   *       *   *       *   *       *   *       *   *       *   *
CGAGCTTTCA ACCCTAAAGA AGTGCTGGTG CGATGGCTGC ATGGAAATGA GGAGCTGTCC CCAGAAAGCT

      1410        1420        1430        1440        1450        1460        1470
       *   *       *   *       *   *       *   *       *   *       *   *       *   *
ACCTAGTGTT TGAGCCCCTA AAGGAGCCAG GCGAGGGAGC CACCACCTAC CTGGTGACAA GCGTGTTGCG

      1480        1490        1500        1510        1520        1530        1540
       *   *       *   *       *   *       *   *       *   *       *   *       *   *
TGTATCAGCT GAAAGCTTGA TATCGAATTC CGGAGGCGGA ACCGGCAGTG CAGCCCGAAG CCCCGCAGTC

      1550        1560        1570        1580        1590
       *   *       *   *       *   *       *   *       *   *
CCCGAGCACG CGTGGCC ATG CGT CCC CTG CGC CCC CGC GCC GCG CTG CTG GCG CTC CTG
                   Met Arg Pro Leu Arg Pro Arg Ala Ala Leu Leu Ala Leu Leu>
                   ___a___a___a___a___a__ORF RF[1] ___a___a___a___a___a___>

      1600        1610        1620        1630        1640        1650
       *   *       *   *       *   *       *   *       *   *       *   *
GCC TCG CTC CTG GCC GCG CCC CCG GTG GCC CCG GCC GAG GCC CCG CAC CTG GTG CAT
Ala Ser Leu Leu Ala Ala Pro Pro Val Ala Pro Ala Glu Ala Pro His Leu Val His>
___a___a___a___a___a___a___a____ORF RF[1] _a___a___a___a___a___a___a___a___>

      1660        1670        1680        1690        1700        1710
       *   *       *   *       *   *       *   *       *   *       *   *
GTG GAC GCG GCC CGC GCG CTG TGG CCC CTG CGG CGC TTC TGG AGG AGC ACA GGC TTC
Val Asp Ala Ala Arg Ala Leu Trp Pro Leu Arg Arg Phe Trp Arg Ser Thr Gly Phe>
___a___a___a___a___a___a___a____ORF RF[1] _a___a___a___a___a___a___a___a___>

      1720        1730        1740        1750        1760        1770
       *   *       *   *       *   *       *   *       *   *       *   *
TGC CCC CCG CTG CCA CAC AGC CAG GCT GAC CAG TAC GTC CTC AGC TGG GAC CAG CAG
Cys Pro Pro Leu Pro His Ser Gln Ala Asp Gln Tyr Val Leu Ser Trp Asp Gln Gln>
___a___a___a___a___a___a___a____ORF RF[1] _a___a___a___a___a___a___a___a___>

      1780        1790        1800        1810        1820
       *   *       *   *       *   *       *   *       *   *
CTC AAC CTC GCC TAT GTG GGC GCC GTC CCT CAC CGC GGC ATC AAG CAG GTC CGG ACC
Leu Asn Leu Ala Tyr Val Gly Ala Val Pro His Arg Gly Ile Lys Gln Val Arg Thr>
___a___a___a___a___a___a___a____ORF RF[1] _a___a___a___a___a___a___a___a___>

      1830        1840        1850        1860        1870        1880
       *   *       *   *       *   *       *   *       *   *       *   *
CAC TGG CTG CTG GAG CTT GTC ACC ACC AGG GGG TCC ACT GGA CGG GGC CTG AGC TAC
His Trp Leu Leu Glu Leu Val Thr Thr Arg Gly Ser Thr Gly Arg Gly Leu Ser Tyr>
```

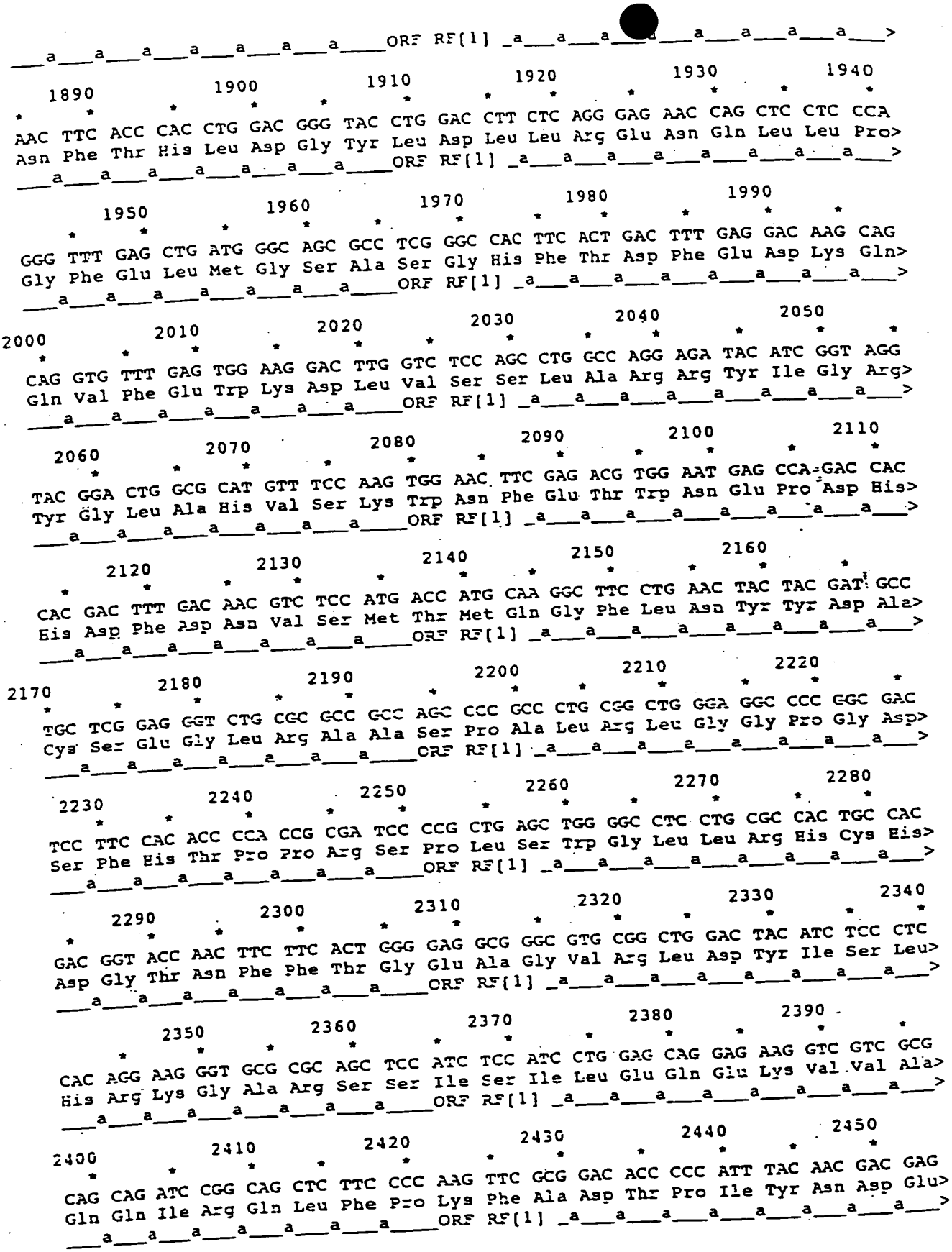
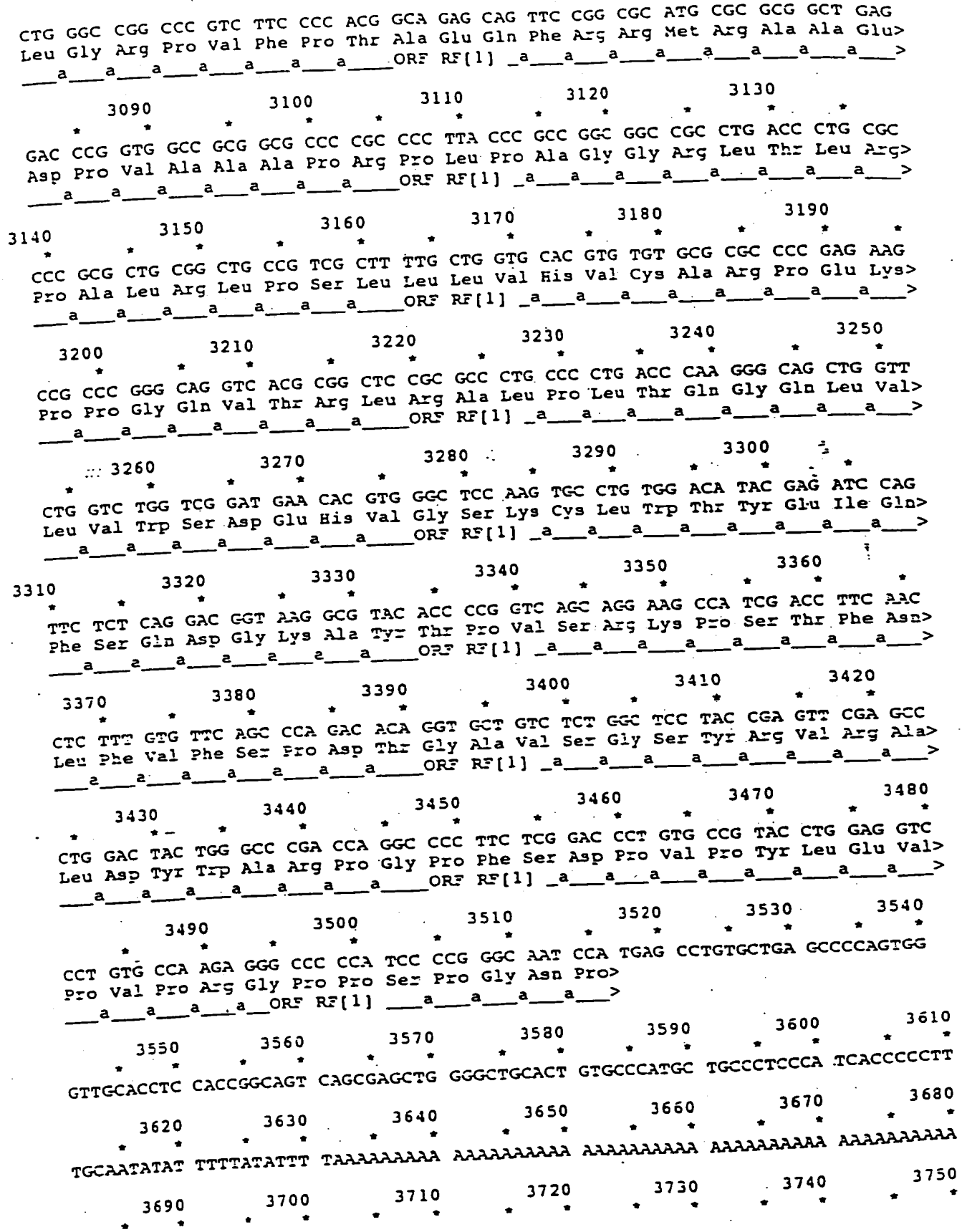

ORF RF[1]
1890 1900 1910 1920 1930 1940
AAC TTC ACC CAC CTG GAC GGG TAC CTG GAC CTT CTC AGG GAG AAC CAG CTC CTC CCA
Asn Phe Thr His Leu Asp Gly Tyr Leu Asp Leu Leu Arg Glu Asn Gln Leu Leu Pro
1950 1960 1970 1980 1990
GGG TTT GAG CTG ATG GGC AGC GCC TCG GGC CAC TTC ACT GAC TTT GAG GAC AAG CAG
Gly Phe Glu Leu Met Gly Ser Ala Ser Gly His Phe Thr Asp Phe Glu Asp Lys Gln
2000 2010 2020 2030 2040 2050
CAG GTG TTT GAG TGG AAG GAC TTG GTC TCC AGC CTG GCC AGG AGA TAC ATC GGT AGG
Gln Val Phe Glu Trp Lys Asp Leu Val Ser Ser Leu Ala Arg Arg Tyr Ile Gly Arg
2060 2070 2080 2090 2100 2110
TAC GGA CTG GCG CAT GTT TCC AAG TGG AAC TTC GAG ACG TGG AAT GAG CCA GAC CAC
Tyr Gly Leu Ala His Val Ser Lys Trp Asn Phe Glu Thr Trp Asn Glu Pro Asp His
2120 2130 2140 2150 2160
CAC GAC TTT GAC AAC GTC TCC ATG ACC ATG CAA GGC TTC CTG AAC TAC TAC GAT GCC
His Asp Phe Asp Asn Val Ser Met Thr Met Gln Gly Phe Leu Asn Tyr Tyr Asp Ala
2170 2180 2190 2200 2210 2220
TGC TCG GAG GGT CTG CGC GCC GCC AGC CCC GCC CTG CGG CTG GGA GGC CCC GGC GAC
Cys Ser Glu Gly Leu Arg Ala Ala Ser Pro Ala Leu Arg Leu Gly Gly Pro Gly Asp
2230 2240 2250 2260 2270 2280
TCC TTC CAC ACC CCA CCG CGA TCC CCG CTG AGC TGG GGC CTC CTG CGC CAC TGC CAC
Ser Phe His Thr Pro Pro Arg Ser Pro Leu Ser Trp Gly Leu Leu Arg His Cys His
2290 2300 2310 2320 2330 2340
GAC GGT ACC AAC TTC TTC ACT GGG GAG GCG GGC GTG CGG CTG GAC TAC ATC TCC CTC
Asp Gly Thr Asn Phe Phe Thr Gly Glu Ala Gly Val Arg Leu Asp Tyr Ile Ser Leu
2350 2360 2370 2380 2390
CAC AGG AAG GGT GCG CGC AGC TCC ATC TCC ATC CTG GAG CAG GAG AAG GTC GTC GCG
His Arg Lys Gly Ala Arg Ser Ser Ile Ser Ile Leu Glu Gln Glu Lys Val Val Ala
2400 2410 2420 2430 2440 2450
CAG CAG ATC CGG CAG CTC TTC CCC AAG TTC GCG GAC ACC CCC ATT TAC AAC GAC GAG
Gln Gln Ile Arg Gln Leu Phe Pro Lys Phe Ala Asp Thr Pro Ile Tyr Asn Asp Glu

FIGURE 1B


2460 2470 2480 2490 2500 2510
GCG GAC CCG CTG GTG GGC TGG TCC CTG CCA CAG CCG TGG AGG GCG GAC GTG ACC TAC
Ala Asp Pro Leu Val Gly Trp Ser Leu Pro Gln Pro Trp Arg Ala Asp Val Thr Tyr
ORF RF[1]
2520 2530 2540 2550 2560
GCG GCC ATG GTG GTG AAG GTC ATC GCG CAG CAT CAG AAC CTG CTA CTG GCC AAC ACC
Ala Ala Met Val Val Lys Val Ile Ala Gln His Gln Asn Leu Leu Leu Ala Asn Thr
ORF RF[1]
2570 2580 2590 2600 2610 2620
ACC TCC GCC TTC CCC TAC GCG CTC CTG AGC AAC GAC AAT GCC TTC CTG AGC TAC CAC
Thr Ser Ala Phe Pro Tyr Ala Leu Leu Ser Asn Asp Asn Ala Phe Leu Ser Tyr His
ORF RF[1]
2630 2640 2650 2660 2670 2680
CCG CAC CCC TTC GCG CAG CGC ACG CTC ACC GCG CGC TTC CAG GTC AAC AAC ACC CGC
Pro His Pro Phe Ala Gln Arg Thr Leu Thr Ala Arg Phe Gln Val Asn Asn Thr Arg
ORF RF[1]
2690 2700 2710 2720 2730
CCG CCG CAC GTG CAG CTG TTG CGC AAG CCG GTG CTC ACG GCC ATG GGG CTG CTG GCG
Pro Pro His Val Gln Leu Leu Arg Lys Pro Val Leu Thr Ala Met Gly Leu Leu Ala
ORF RF[1]
2740 2750 2760 2770 2780 2790
CTG CTG GAT GAG GAG CAG CTC TGG GCC GAA GTG TCG CAG GCC GGG ACC GTC CTG GAC
Leu Leu Asp Glu Glu Gln Leu Trp Ala Glu Val Ser Gln Ala Gly Thr Val Leu Asp
ORF RF[1]
2800 2810 2820 2830 2840 2850
AGC AAC CAC ACG GTG GGC GTC CTG GCC AGC GCC CAC CGC CCC CAG GGC CCG GCC GAC
Ser Asn His Thr Val Gly Val Leu Ala Ser Ala His Arg Pro Gln Gly Pro Ala Asp
ORF RF[1]
2860 2870 2880 2890 2900 2910
GCC TGG CGC GCC GCG GTG CTG ATC TAC GCG AGC GAC GAC ACC CGC GCC CAC CCC AAC
Ala Trp Arg Ala Ala Val Leu Ile Tyr Ala Ser Asp Asp Thr Arg Ala His Pro Asn
ORF RF[1]
2920 2930 2940 2950 2960
CGC AGC GTC GCG GTG ACC CTG CGG CTG CGC GGG GTG CCC CCC GGC CCG GGC CTG GTC
Arg Ser Val Ala Val Thr Leu Arg Leu Arg Gly Val Pro Pro Gly Pro Gly Leu Val
ORF RF[1]
2970 2980 2990 3000 3010 3020
TAC GTC ACG CGC TAC CTG GAC AAC GGG CTC TGC AGC CCC GAC GGC GAG TGG CGG CGC
Tyr Val Thr Arg Tyr Leu Asp Asn Gly Leu Cys Ser Pro Asp Gly Glu Trp Arg Arg
ORF RF[1]
3030 3040 3050 3060 3070 3080


CTG GGC CGG CCC GTC TTC CCC ACG GCA GAG CAG TTC CGG CGC ATG CGC GCG GCT GAG
Leu Gly Arg Pro Val Phe Pro Thr Ala Glu Gln Phe Arg Arg Met Arg Ala Ala Glu>
ORF RF[1]
3090 3100 3110 3120 3130
GAC CCG GTG GCC GCG GCG CCC CGC CCC TTA CCC GCC GGC GGC CGC CTG ACC CTG CGC
Asp Pro Val Ala Ala Ala Pro Arg Pro Leu Pro Ala Gly Gly Arg Leu Thr Leu Arg>
ORF RF[1]
3140 3150 3160 3170 3180 3190
CCC GCG CTG CGG CTG CCG TCG CTT TTG CTG GTG CAC GTG TGT GCG CGC CCC GAG AAG
Pro Ala Leu Arg Leu Pro Ser Leu Leu Leu Val His Val Cys Ala Arg Pro Glu Lys>
ORF RF[1]
3200 3210 3220 3230 3240 3250
CCG CCC GGG CAG GTC ACG CGG CTC CGC GCC CTG CCC CTG ACC CAA GGG CAG CTG GTT
Pro Pro Gly Gln Val Thr Arg Leu Arg Ala Leu Pro Leu Thr Gln Gly Gln Leu Val>
ORF RF[1]
3260 3270 3280 3290 3300
CTG GTC TGG TCG GAT GAA CAC GTG GGC TCC AAG TGC CTG TGG ACA TAC GAG ATC CAG
Leu Val Trp Ser Asp Glu His Val Gly Ser Lys Cys Leu Trp Thr Tyr Glu Ile Gln>
ORF RF[1]
3310 3320 3330 3340 3350 3360
TTC TCT CAG GAC GGT AAG GCG TAC ACC CCG GTC AGC AGG AAG CCA TCG ACC TTC AAC
Phe Ser Gln Asp Gly Lys Ala Tyr Thr Pro Val Ser Arg Lys Pro Ser Thr Phe Asn>
ORF RF[1]
3370 3380 3390 3400 3410 3420
CTC TTT GTG TTC AGC CCA GAC ACA GGT GCT GTC TCT GGC TCC TAC CGA GTT CGA GCC
Leu Phe Val Phe Ser Pro Asp Thr Gly Ala Val Ser Gly Ser Tyr Arg Val Arg Ala>
ORF RF[1]
3430 3440 3450 3460 3470 3480
CTG GAC TAC TGG GCC CGA CCA GGC CCC TTC TCG GAC CCT GTG CCG TAC CTG GAG GTC
Leu Asp Tyr Trp Ala Arg Pro Gly Pro Phe Ser Asp Pro Val Pro Tyr Leu Glu Val>
ORF RF[1]
3490 3500 3510 3520 3530 3540
CCT GTG CCA AGA GGG CCC CCA TCC CCG GGC AAT CCA TGAG CCTGTGCTGA GCCCCAGTGG
Pro Val Pro Arg Gly Pro Pro Ser Pro Gly Asn Pro>
ORF RF[1]
3550 3560 3570 3580 3590 3600 3610
GTTGCACCTC CACCGGCAGT CAGCGAGCTG GGGCTGCACT GTGCCCATGC TGCCCTCCCA TCACCCCCTT
3620 3630 3640 3650 3660 3670 3680
TGCAATATAT TTTTATATTT TAAAAAAAAA AAAAAAAAAA AAAAAAAAAA AAAAAAAAAA AAAAAAAAAA
3690 3700 3710 3720 3730 3740 3750

AAAAAAAAAA AAAAAAAAAG AATTCCTGCA GCCCGGGGGA TCCACTAGTT CTAGAGGGCC CGTTTAAACC

3760 3770 3780 3790 3800 3810 3820

CGCTGATCAG CCTCGACTGT GCCTTCTAGT TGCCAGCCAT CTGTTGTTTG CCCCTCCCCC GTGCCTTCCT

3830 3840 3850 3860 3870 3880 3890

TGACCCTGGA AGGTGCCACT CCCACTGTCC TTTCCTAATA AAATGAGGAA ATTGCATCGC ATTGTCTGAG

3900 3910 3920 3930 3940 3950 3960

TAGGTGTCAT TCTATTCTGG GGGGTGGGGT GGGGCAGGAC AGCAAGGGGG AGGATTGGGA AGACAATAGC

3970 3980 3990 4000 4010 4020 4030

AGGCATGCTG GGGATGCGGT GGGCTCTATG GCTTCTGAGG CGGAAAGAAC CAGCTGGGGC TCGAGAGCTT

4040 4050 4060 4070 4080 4090 4100

GGCGTAATCA TGGTCATAGC TGTTTCCTGT GTGAAATTGT TATCCGCTCA CAATTCCACA CAACATACGA

4110 4120 4130 4140 4150 4160 4170

GCCGGAAGCA TAAAGTGTAA AGCCTGGGGT GCCTAATGAG TGAGCTAACT CACATTAATT GCGTTGCGCT

4180 4190 4200 4210 4220 4230 4240

CACTGCCCGC TTTCCAGTCG GGAAACCTGT CGTGCCAGCT GCATTAATGA ATCGGCCAAC GCGCGGGGAG

4250 4260 4270 4280 4290 4300 4310

AGGCGGTTTG CGTATTGGGC GCTCTTCCGC TTCCTCGCTC ACTGACTCGC TGCGCTCGGT CGTTCGGCTG

4320 4330 4340 4350 4360 4370 4380

CGGCGAGCGG TATCAGCTCA CTCAAAGGCG GTAATACGGT TATCCACAGA ATCAGGGGAT AACGCAGGAA

4390 4400 4410 4420 4430 4440 4450

AGAACATGTG AGCAAAAGGC CAGCAAAAGG CCAGGAACCG TAAAAAGGCC GCGTTGCTGG CGTTTTTCCA

4460 4470 4480 4490 4500 4510 4520

TAGGCTCCGC CCCCCTGACG AGCATCACAA AAATCGACGC TCAAGTCAGA GGTGGCGAAA CCCGACAGGA

4530 4540 4550 4560 4570 4580 4590

CTATAAAGAT ACCAGGCGTT TCCCCCTGGA AGCTCCCTCG TGCGCTCTCC TGTTCCGACC CTGCCGCTTA

4600 4610 4620 4630 4640 4650 4660

CCGGATACCT GTCCGCCTTT CTCCCTTCGG GAAGCGTGGC GCTTTCTCAA TGCTCACGCT GTAGGTATCT

4670 4680 4690 4700 4710 4720 4730

CAGTTCGGTG TAGGTCGTTC GCTCCAAGCT GGGCTGTGTG CACGAACCCC CCGTTCAGCC CGACCGCTGC

4740 4750 4760 4770 4780 4790 4800

GCCTTATCCG GTAACTATCG TCTTGAGTCC AACCCGGTAA GACACGACTT ATCGCCACTG GCAGCAGCCA

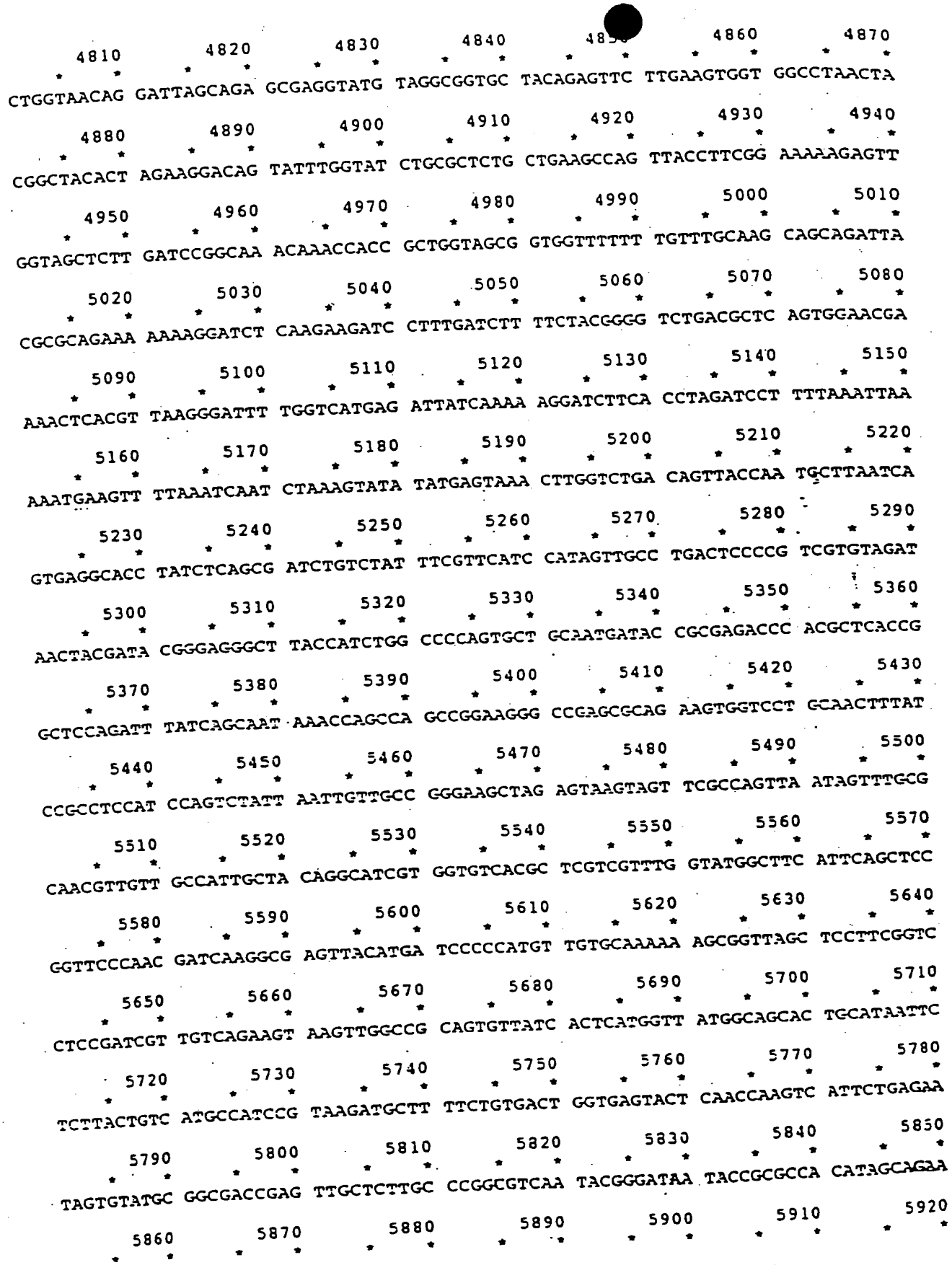

4810 4820 4830 4840 4860 4870
CTGGTAACAG GATTAGCAGA GCGAGGTATG TAGGCGGTGC TACAGAGTTC TTGAAGTGGT GGCCTAACTA
4880 4890 4900 4910 4920 4930 4940
CGGCTACACT AGAAGGACAG TATTTGGTAT CTGCGCTCTG CTGAAGCCAG TTACCTTCGG AAAAAGAGTT
4950 4960 4970 4980 4990 5000 5010
GGTAGCTCTT GATCCGGCAA ACAAACCACC GCTGGTAGCG GTGGTTTTTT TGTTTGCAAG CAGCAGATTA
5020 5030 5040 5050 5060 5070 5080
CGCGCAGAAA AAAAGGATCT CAAGAAGATC CTTTGATCTT TTCTACGGGG TCTGACGCTC AGTGGAACGA
5090 5100 5110 5120 5130 5140 5150
AAACTCACGT TAAGGGATTT TGGTCATGAG ATTATCAAAA AGGATCTTCA CCTAGATCCT TTTAAATTAA
5160 5170 5180 5190 5200 5210 5220
AAATGAAGTT TTAAATCAAT CTAAAGTATA TATGAGTAAA CTTGGTCTGA CAGTTACCAA TGCTTAATCA
5230 5240 5250 5260 5270 5280 5290
GTGAGGCACC TATCTCAGCG ATCTGTCTAT TTCGTTCATC CATAGTTGCC TGACTCCCCG TCGTGTAGAT
5300 5310 5320 5330 5340 5350 5360
AACTACGATA CGGGAGGGCT TACCATCTGG CCCCAGTGCT GCAATGATAC CGCGAGACCC ACGCTCACCG
5370 5380 5390 5400 5410 5420 5430
GCTCCAGATT TATCAGCAAT AAACCAGCCA GCCGGAAGGG CCGAGCGCAG AAGTGGTCCT GCAACTTTAT
5440 5450 5460 5470 5480 5490 5500
CCGCCTCCAT CCAGTCTATT AATTGTTGCC GGGAAGCTAG AGTAAGTAGT TCGCCAGTTA ATAGTTTGCG
5510 5520 5530 5540 5550 5560 5570
CAACGTTGTT GCCATTGCTA CAGGCATCGT GGTGTCACGC TCGTCGTTTG GTATGGCTTC ATTCAGCTCC
5580 5590 5600 5610 5620 5630 5640
GGTTCCCAAC GATCAAGGCG AGTTACATGA TCCCCCATGT TGTGCAAAAA AGCGGTTAGC TCCTTCGGTC
5650 5660 5670 5680 5690 5700 5710
CTCCGATCGT TGTCAGAAGT AAGTTGGCCG CAGTGTTATC ACTCATGGTT ATGGCAGCAC TGCATAATTC
5720 5730 5740 5750 5760 5770 5780
TCTTACTGTC ATGCCATCCG TAAGATGCTT TTCTGTGACT GGTGAGTACT CAACCAAGTC ATTCTGAGAA
5790 5800 5810 5820 5830 5840 5850
TAGTGTATGC GGCGACCGAG TTGCTCTTGC CCGGCGTCAA TACGGGATAA TACCGCGCCA CATAGCAGAA
5860 5870 5880 5890 5900 5910 5920

FIGURE 15

CTTTAAAAGT GCTCATCATT GGAAAACGTT CTTCGGGGCG AAAACTCTCA AGGATCTTAC CGCTGTTGAG

5930 5940 5950 5960 5970 5980 5990

ATCCAGTTCG ATGTAACCCA CTCGTGCACC CAACTGATCT TCAGCATCTT TTACTTTCAC CAGCGTTTCT

6000 6010 6020 6030 6040 6050 6060

GGGTGAGCAA AAACAGGAAG GCAAAATGCC GCAAAAAAGG GAATAAGGGC GACACGGAAA TGTTGAATAC

6070 6080 6090 6100 6110 6120 6130

TCATACTCTT CCTTTTTCAA TATTATTGAA GCATTTATCA GGGTTATTGT CTCATGAGCG GATACATATT

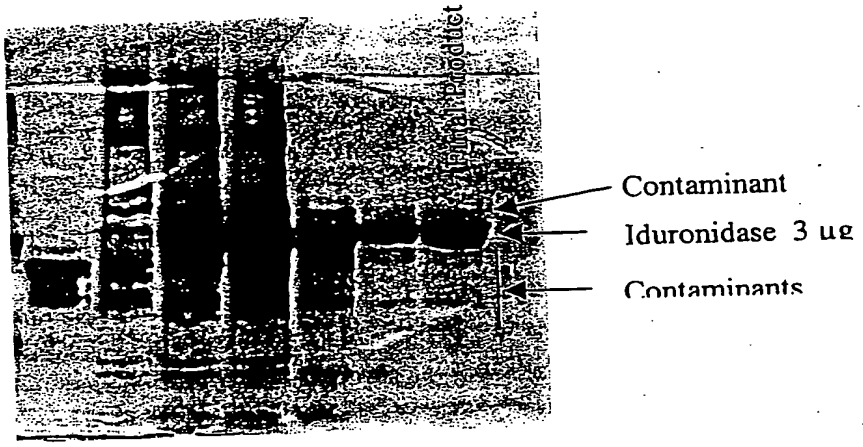
6140 6150 6160 6170 6180 6190 6200

TGAATGTATT TAGAAAAATA AACAAATAGG GGTTCCGCGC ACATTTCCCC GAAAAGTGCC ACCTGACGTC

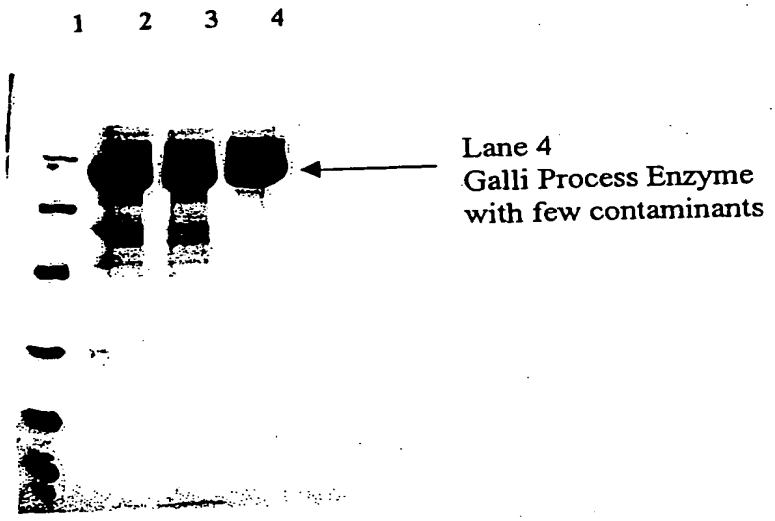
FIGURE 1G

## FIGURE 2. SDS-POLYACRYLAMIDE GELS DEMONSTRATING IMPROVEMENTS IN PURITY

Gel using the Kakkis et al 1994, published procedure for purification




Gel using the new Galli Process contained in this application




1. Molecular Weight Marker
2. Prior Process Carson (nonpublished) Batch 2000C9001 Reference Reduced (7.5 µg)
3. Same Batch 2000C9001 Reference Reduced (5.0 µg)
4. Galli Process Enzyme Batch P10006 (5.0 µg)

FIGURE 2

FIGURE 3A IDURONIDASE PRODUCTION USING THE GALLI PROCESS

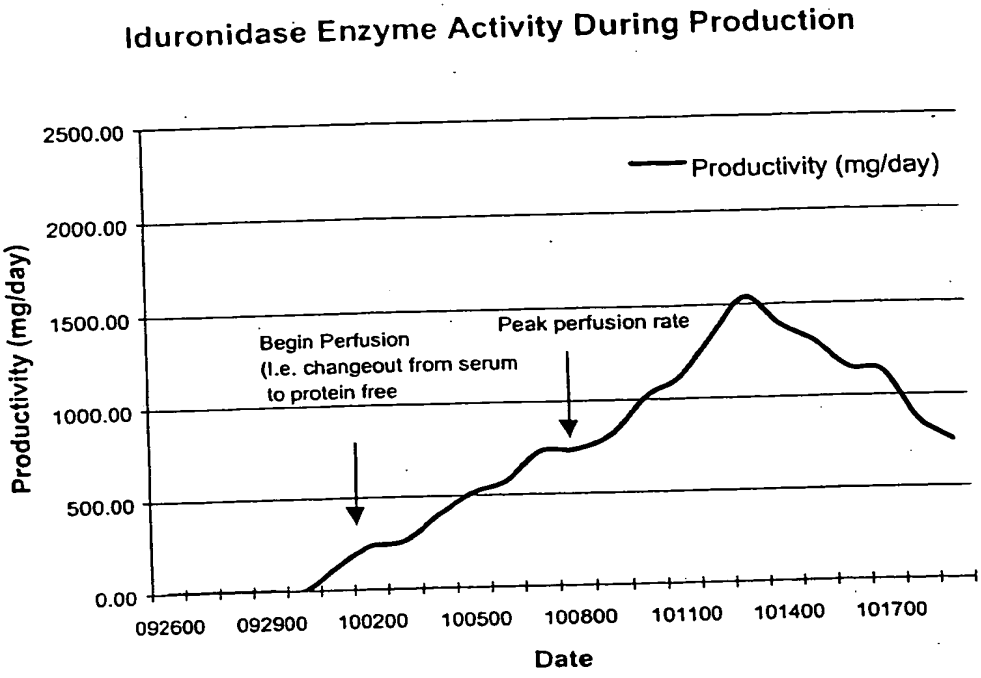
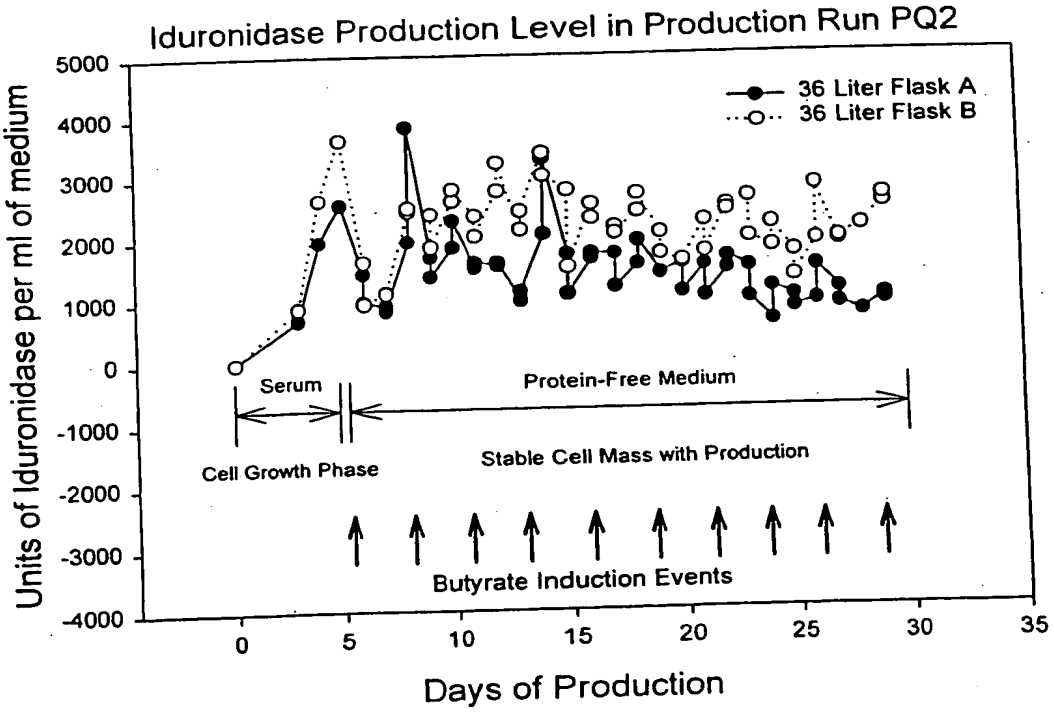
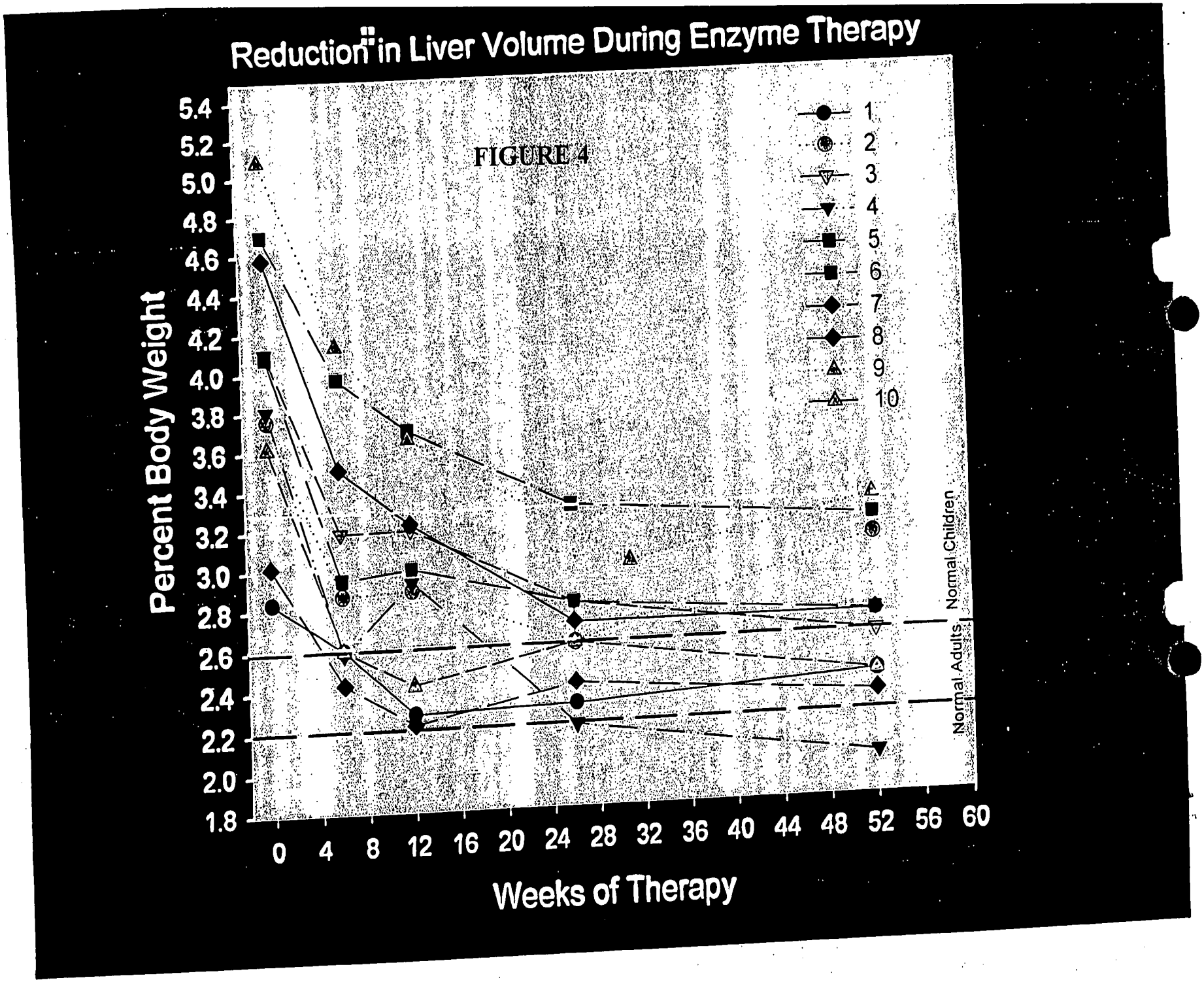
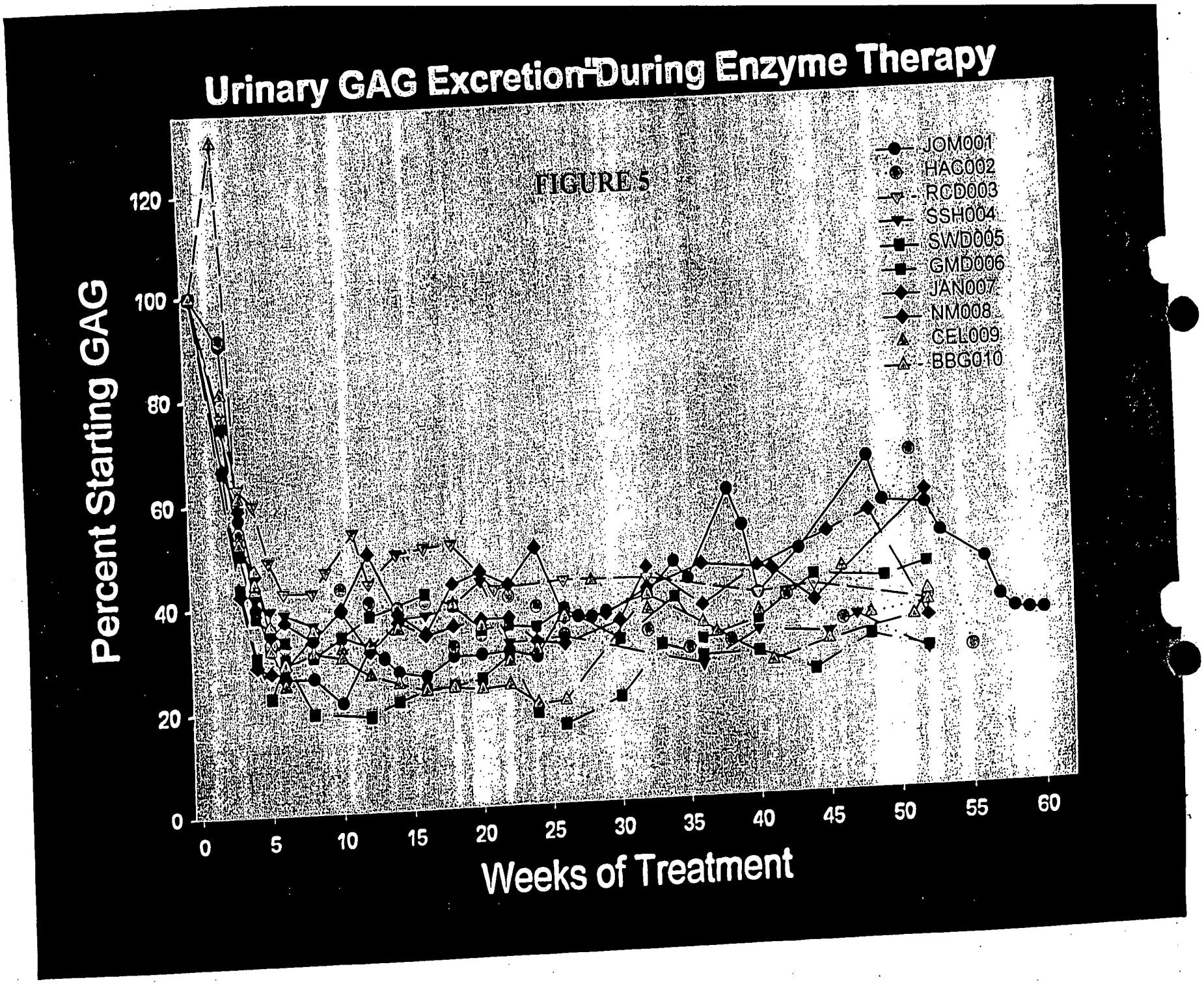
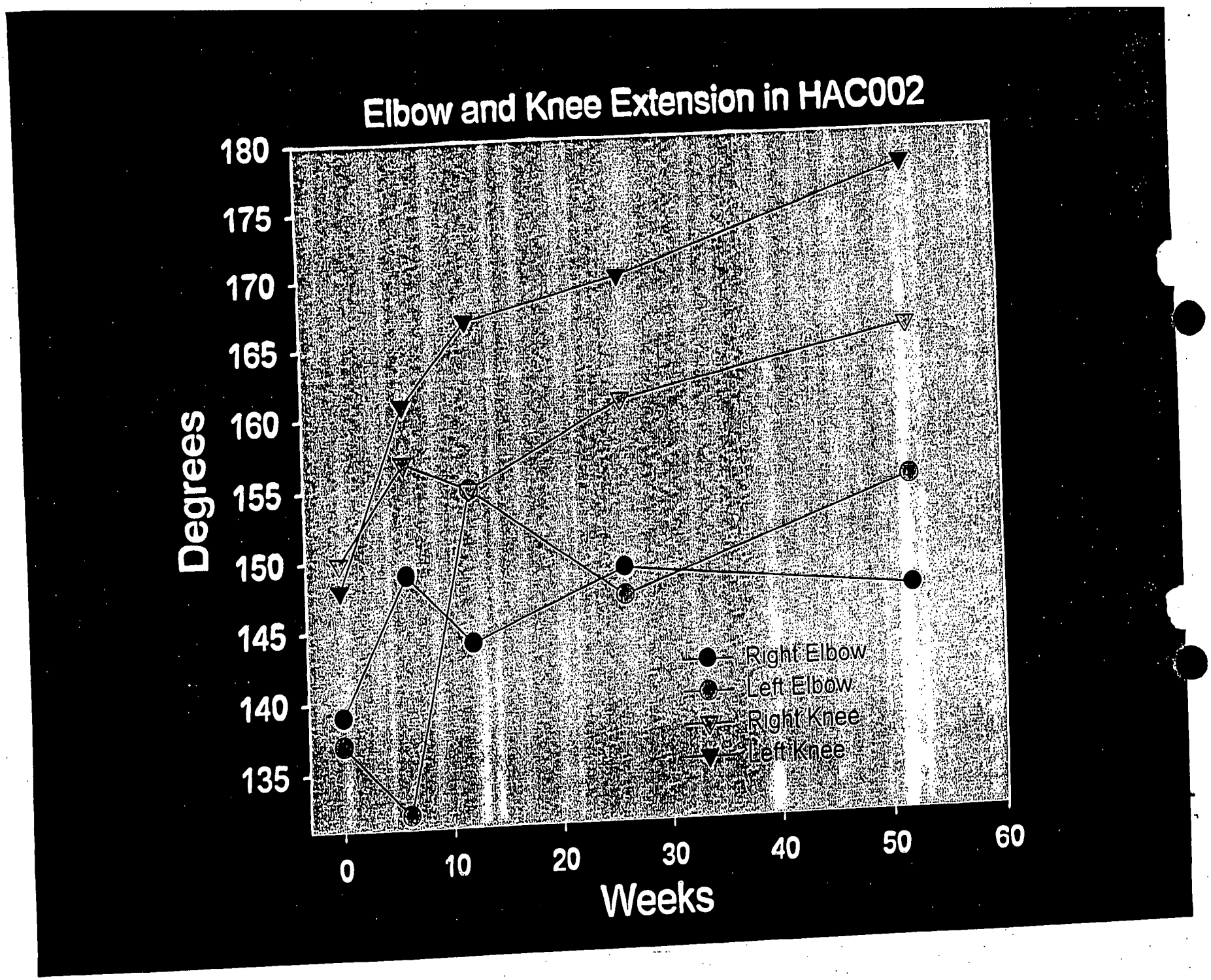


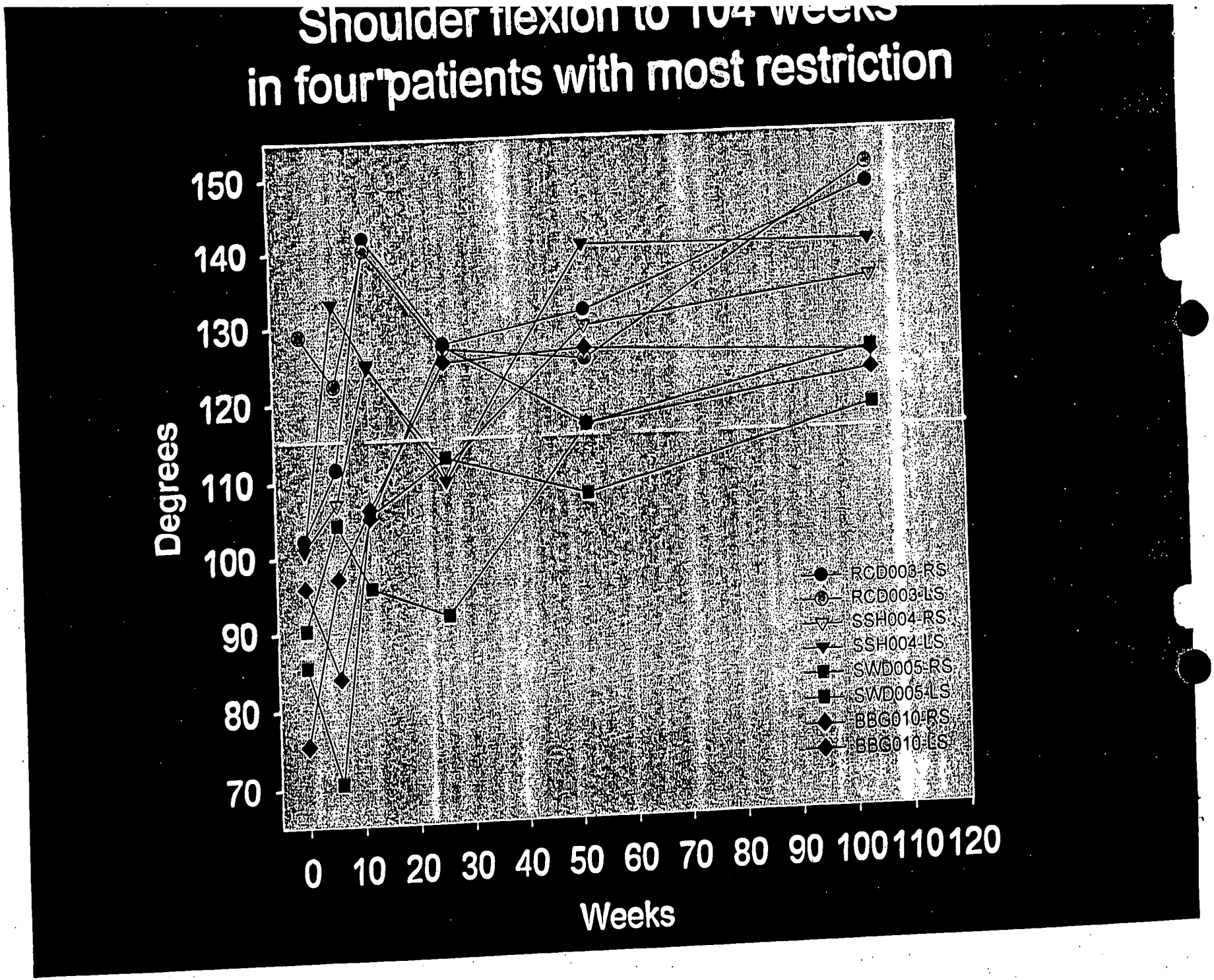

FIGURE 3B. IDURONIDASE PRODUCTION USING BUTYRATE INDUCTION




Reduction in Liver Volume During Enzyme Therapy
FIGURE 4
Percent Body Weight
5.4
5.2
5.0
4.8
4.6
4.4
4.2
4.0
3.8
3.6
3.4
3.2
3.0
2.8
2.6
2.4
2.2
2.0
1.8
0
4
8
12
16
20
24
28
32
36
40
44
48
52
56
60
Weeks of Therapy
1
2
3
4
5
6
7
8
9
10
Normal Children
Normal Adults

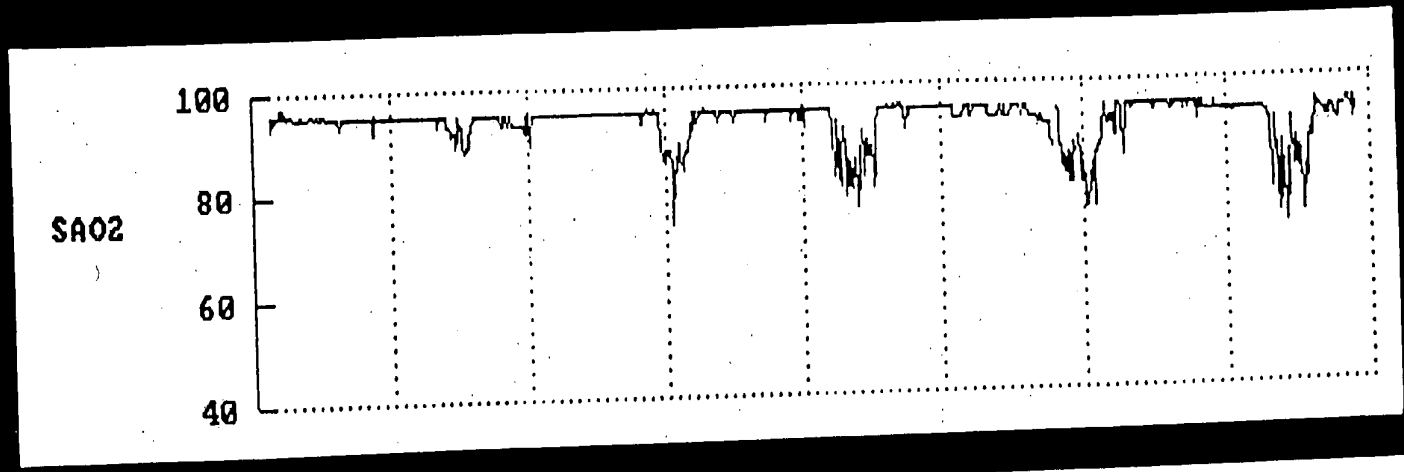

Urinary GAG Excretion During Enzyme Therapy
FIGURE 5
Percent Starting GAG
Weeks of Treatment
0
20
40
60
80
100
120
0
5
10
15
20
25
30
35
40
45
50
55
60
JOM001
HAC002
RCD003
SSH004
SWD005
GMD006
JAN007
NM008
CEL009
BBG010


Elbow and Knee Extension in HAC002
180
175
170
165
160
155
150
145
140
135
Degrees
0
10
20
30
40
50
60
Weeks
Right Elbow
Left Elbow
Right Knee
Left Knee

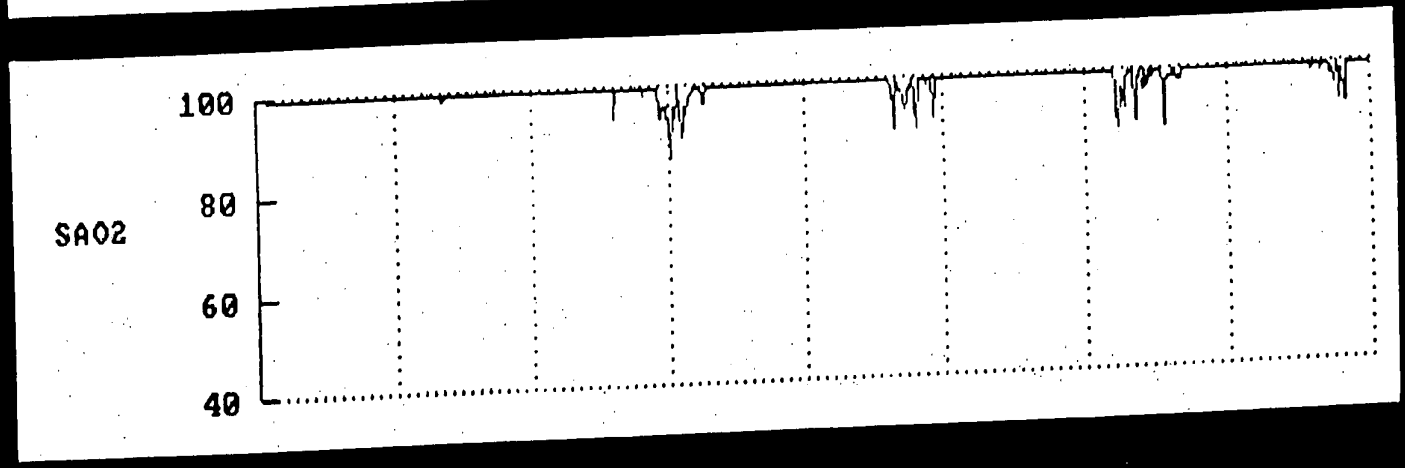

Shoulder flexion to 104 weeks
in four patients with most restriction
150
140
130
120
110
100
90
80
70
Degrees
0
10
20
30
40
50
60
70
80
90
100
110
120
Weeks
RCD003-RS
RCD003-LS
SSH004-RS
SSH004-LS
SWD005-RS
SWD005-LS
BBG010-RS
BBG010-LS

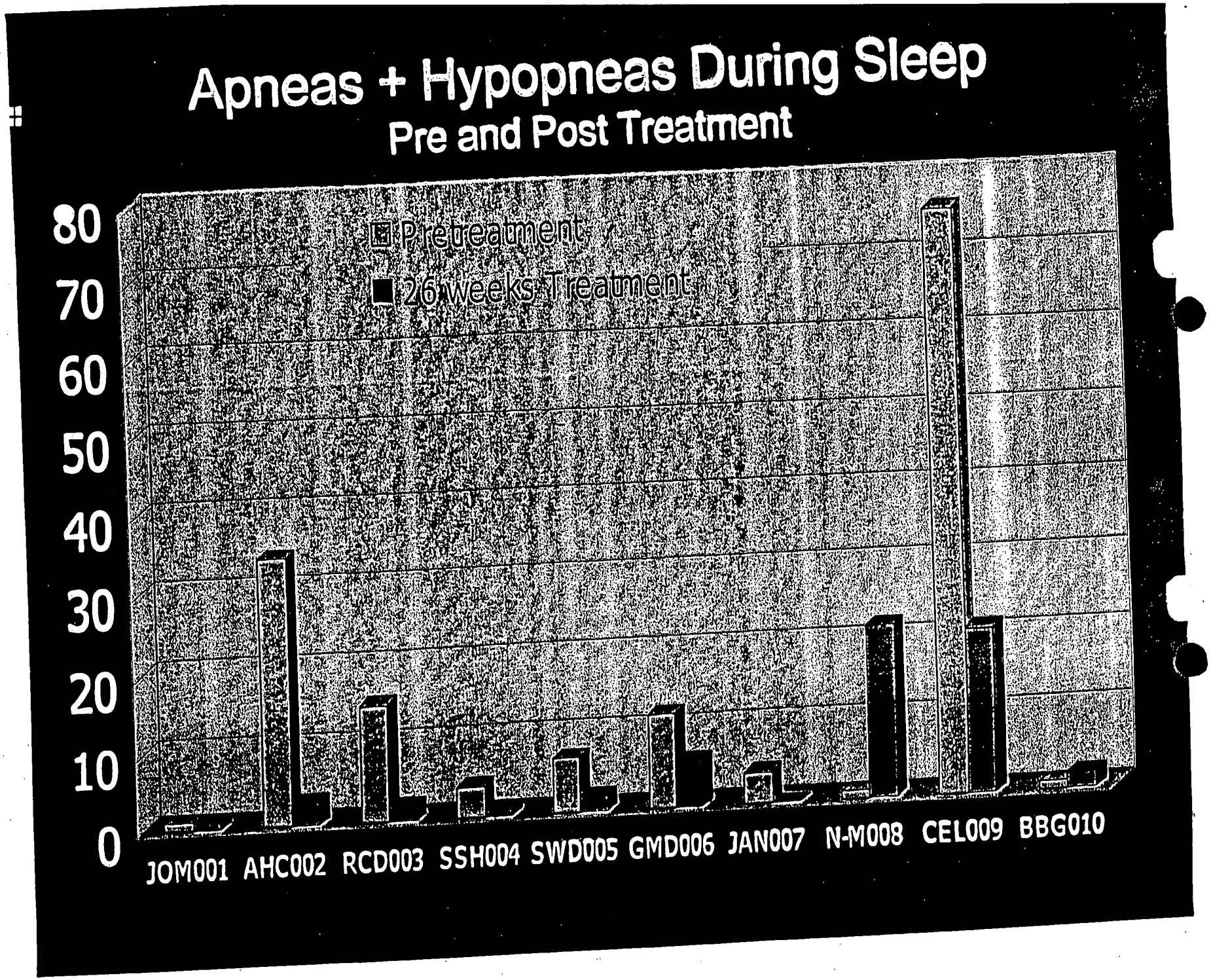
# Sleep Apnea Improves

0 weeks

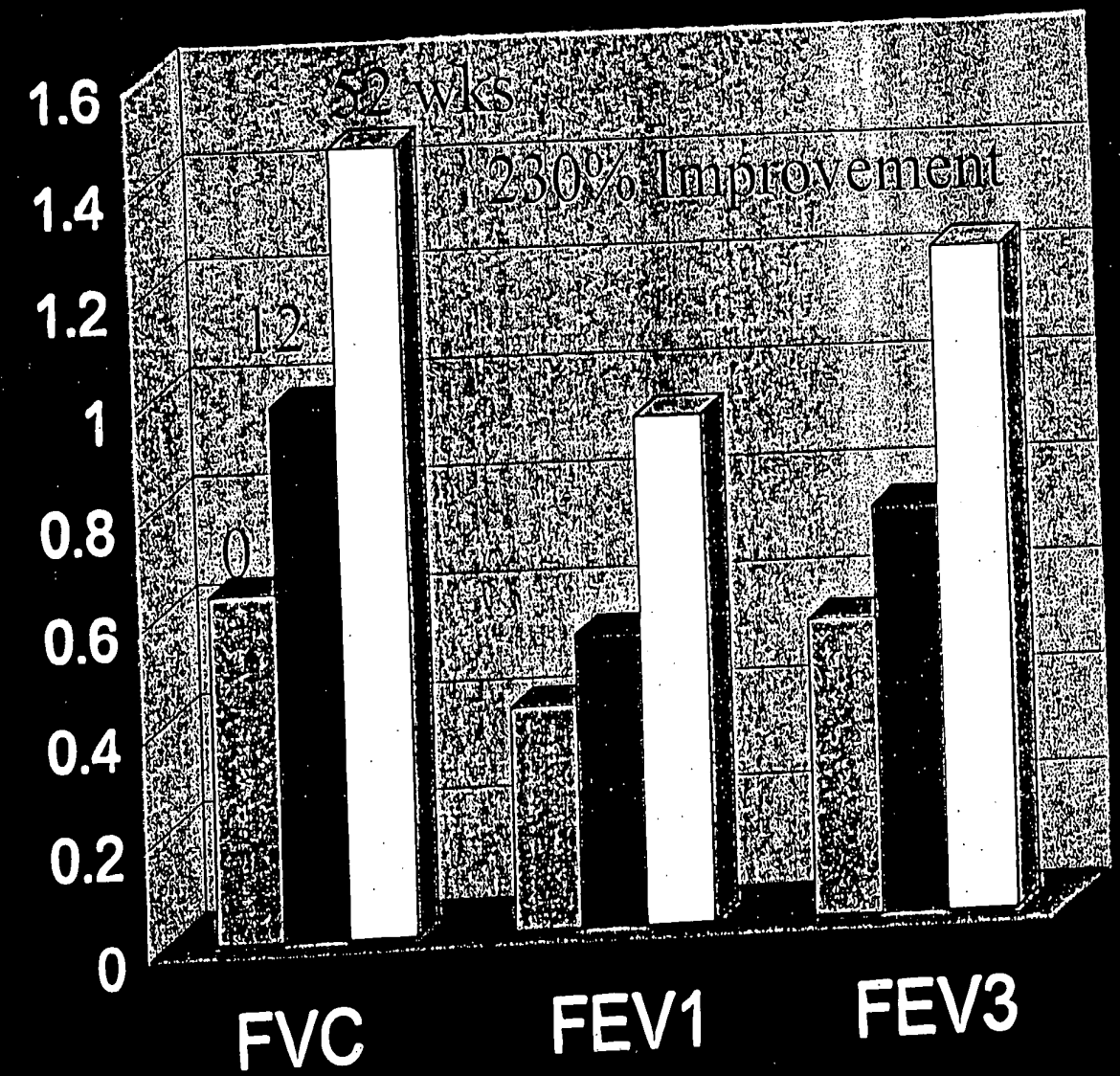



After
6 weeks




Apneas + Hypopneas During Sleep
Pre and Post Treatment
Pretreatment
26 weeks Treatment
80
70
60
50
40
30
20
10
0
JOM001
AHC002
RCD003
SSH004
SWD005
GMD006
JAN007
N-M008
CEL009
BBG010

# Pulmonary Function Tests in GMD006



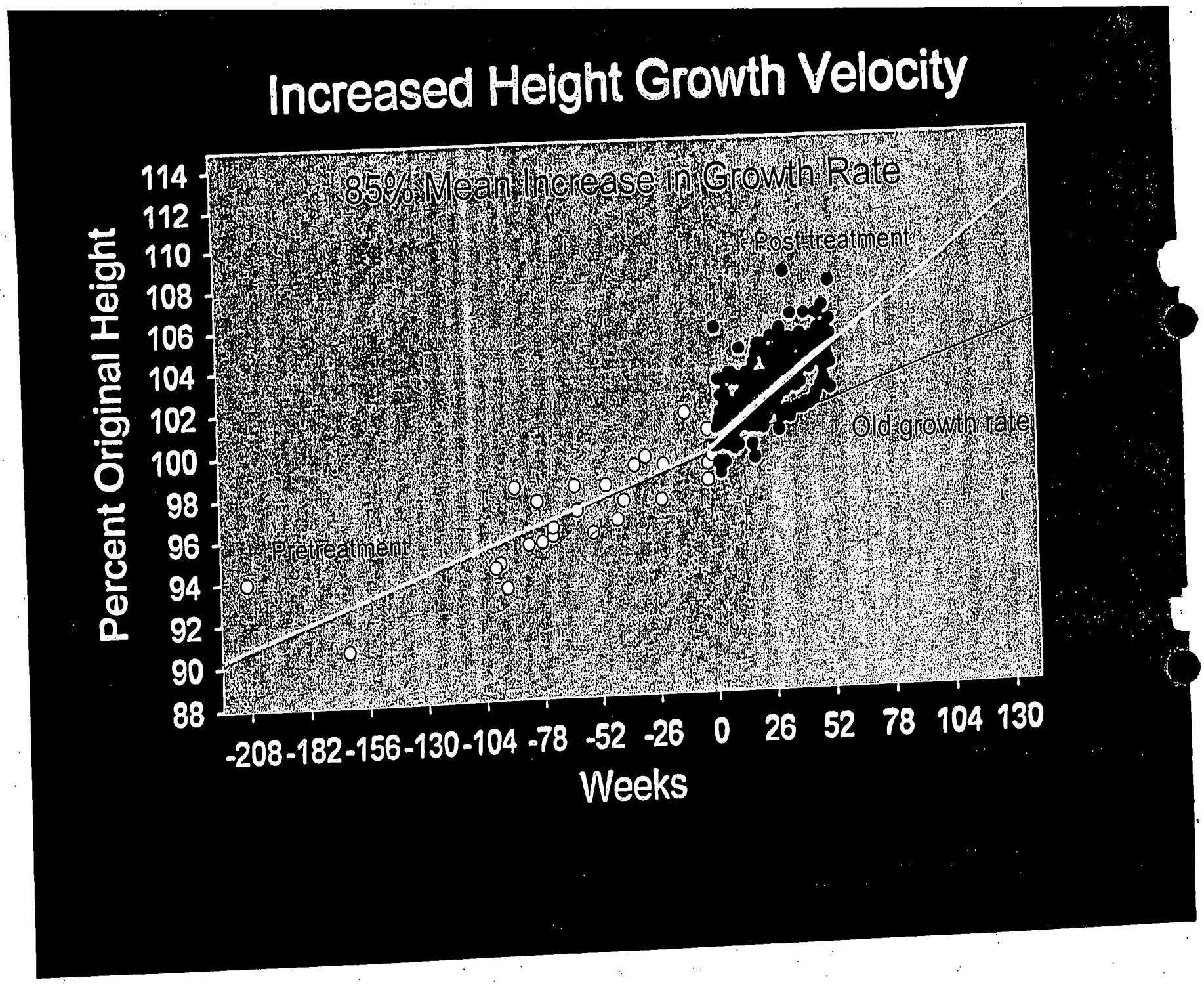

Increased Height Growth Velocity
85% Mean Increase in Growth Rate
Post-treatment
Old growth rate
Pretreatment
Percent Original Height
114
112
110
108
106
104
102
100
98
96
94
92
90
88
-208 -182 -156 -130 -104 -78 -52 -26 0 26 52 78 104 130
Weeks

FIGURE 12.

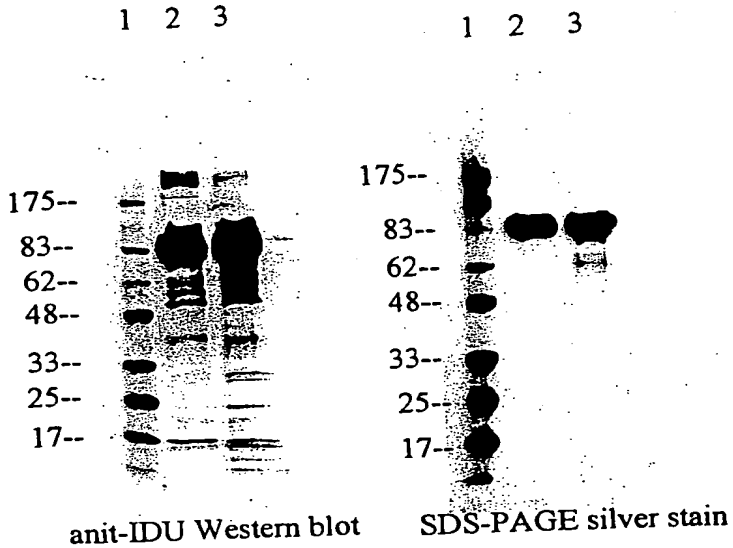
COMPARISON OF HOST PROTEIN CONTAMINATION BETWEEN A PRIOR AND THE NEW GALLI PROCESS

Chinese Hamster Ovary Host Protein Contamination by ELISA Assay

| SOURCE AND BATCH NUMBER | CHOP PROTEIN CONTAMINATION (microgram per milligram) | PERCENT CHOP CONTAMINATION | PURITY OF THE ENZYME FROM CHOP |
|---|---|---|---|
| Prior Process (Carson/REI) | | | |
| C9002 | 14 | 1.4% | 98.6% |
| C9003 | 24 | 2.4% | 97.6% |
| C9004 | 16 | 1.6% | 98.4% |
| New Process (Galli) | | | |
| P1003 | <1.3 | <0.13% | >99.9% |
| P1006 | 1.2 | 0.12% | 99.9% |
| P1007 | <0.6 | <0.06% | >99.9% |
| P1008 | <0.67 | <0.067% | >99.9% |

FIGURE 12

## Comparison of Galli and Carson Material




anit-IDU Western blot
1:50,000

SDS-PAGE silver stain

1 Marker
2 Galli Referenced-0201
3 Carson C9002

5ug/lane

FIGURE 13